LEUCOSTASIS: AN UNCOMMON CAUSE OF RESPIRATORY DISTRESS

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INTRODUCTION

Leucostasis occasionally develops in patients with Leukemia. In this condition there is sludging of the capillary circulation due to large numbers of leukemic cells. The organs frequently affected are the central nervous system and the lungs. Leucostasis occurs mainly in acute leukemias and in acute blast crisis of chronic myeloid leukemia. Respiratory involvement manifests as reversible respiratory distress and we report a patient presenting as such.

CASE REPORT

A 26-year-old Indian man, presented with lethargy and a progressively enlarging left hypochondrial mass for one year. There was associated anorexia and weight loss. Two weeks before admission he developed exertional dyspnoea which progressed to dyspnoea at rest. There was cough productive of scanty whitish sputum but he had no orthopnoea, parosyxmal nocturnal dyspnoea or ankle edema.

On admission, he was tachypnoeic and cyanosed. His pulse rate was 100/minute and his blood pressure was 110/80 mm Hg. There were no obvious bleeding manifestations. The liver was enlarged to 10 cms, below the (R) costal margin; the spleen to 14 cms below the left costal edge.

Haemotological investigations showed a Haemoglobin of 6.0 g/dl, platelets count of 200,000/mm^3 and a total white count of 760,000/mm^3 with a differential count of 46% neutrophils, 2% lymphocytes, 32% metamyelocytes, 18% myelocytes and 2% blast cells. The peripheral blood picture was consistent with Chronic Myeloid Leukemia.

Bone marrow could not be aspirated. A trephine biopsy showed marked granulocytic hyperplasia and a shift of white count to the left. The marrow findings confirmed Chronic Myeloid Leukemia.

Arterial blood gas analysis showed marked hypoxia with a PO2 of 3.9 kPa and a slightly elevated PCO2 of 6.46 kPa.

There were diffuse, patchy opacities in both lung fields in the Chest X-ray (Fig. 1). Repeated sputum smears and cultures showed no organisms. Lung volumes and tests of ventilatory functions were within normal limits.

In view of the extremely high white cell count, cyclophosphamide 1 gm was administered intravenously, followed by oral Busulfan and Thioguanine. No antibiotics or diuretics were given.

With this treatment, the white cell count dropped to less than 600,000/mm^3 within one week and to less than 300,000/mm^3 by the end of three weeks, accompanied by marked improvement in his clinical condition.

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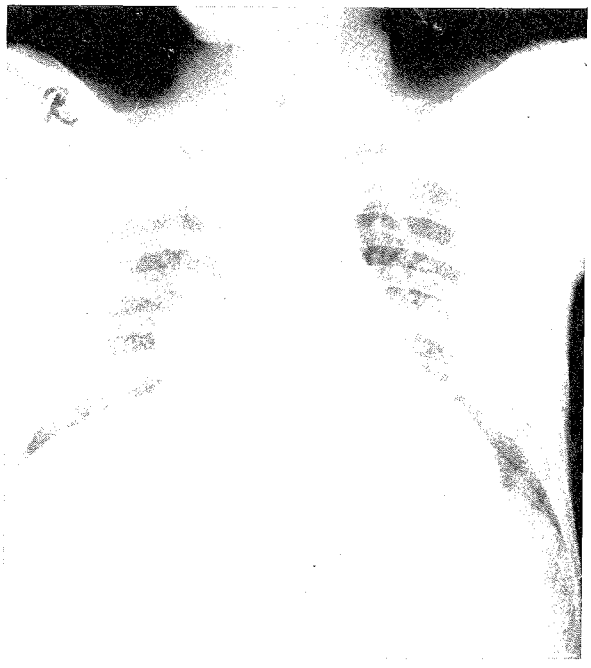

Fig. 1 Lungs showing patchy opacities.

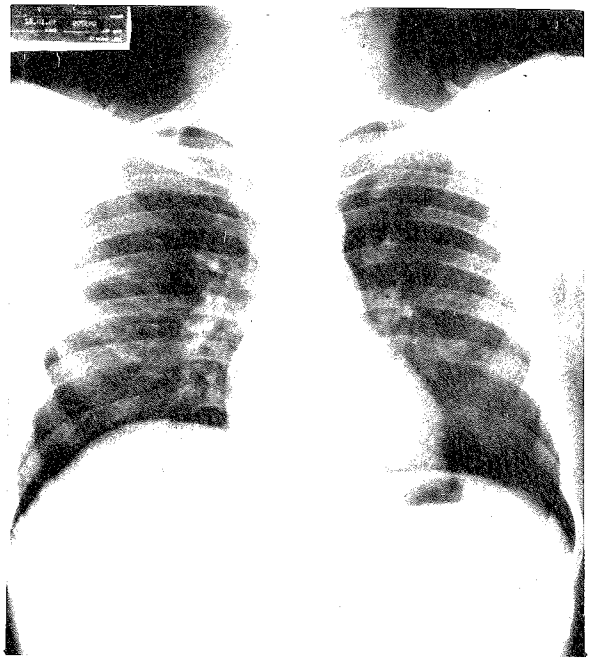

Fig. 2 Chest X-ray after treatment.

Eight weeks after admission, he was able to perform normal physical tasks without being breathless. (His appetite was good and he had put on weight). The spleen had shrunk to half of its original size, haemoglobin was 10.9 g/dl and the total white count was 29,000 with a differential of 84% neutrophils, 4% myelocytes and 8% metamyelocytes. Arterial blood gas showed a PO2 of 11.0 kPa and a PCO2 of 5.0 kPa. The Chest X-ray was normal. (Fig. 2).

DISCUSSION

In leukemia, major respiratory problems are most often due to infections, haemorrhage, cardiac failure, drug therapy or pulmonary edema of various causes.

In this patient, the correlation of severe respiratory symptoms with an extremely high total white count and the improvement noted, with a decrease in the total white count suggests that the respiratory distress was due to leucostasis.

One of the earliest records of leucostasis in the lungs was by Jaochim and Loewe. [1] They presented a case of Acute Myeloid Leukemia characterised by migratory infiltration of both lungs which were ascribed to "repeated infarctions" secondary to vascular occlusion with myeloid cells. Fiesenger and Fauvet [2] attached similar significance to vascular occlusion. In a patient who succumbed to Chronic Myeloid Leukemia, they found numerous vessels 3 or 4 mm in size completely occluded by white thrombi made up of leukemic cells.

Many reports of leucostasis in the lungs have since appeared in the literature. [3,4]

In this clinical syndrome several features have been noted. Vernant *et al* felt that the rapidity of increase of the total white count was important in the development of respiratory distress. Leukemic patients who had high white cell count with a high percentage of blasts but whose counts were stable developed no respiratory symptoms. Also the reduction of the total white count was correlated with an improvement in symptoms in all their patients.

The higher frequency of leucostasis in Acute Myeloid Leukemia, as compared to other forms of leukemia is probably due to the lower degree of cellular deformability in the former. Lichtman [5] found myeloblasts to be the least deformable; as they mature the white cells become more deformable.

The predominance of leucostasis in the lungs and central nervous systems remains unexplained, possibly being the consequence of a particular local microcirculation.

Finally, urgent treatment to reduce the white cells count is important. With the availability of cell-separators, leukopharesis is probably the treatment of choice.

ACKNOWLEDGEMENT

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